Familial Mediterranean Fever Related Spondyloarthritis

Ailesel Akdeniz Ateşi İlişkili Spondiloartrit

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Özet

Ailesel Akdeniz Ateşi (AAA) febril serozit atakları ile karakterize bir otozomal resesif hastalıktır. En sık görülen semptom abdominal ve ikincisi artiküler ataktır. Eklem tutulumu genellikle alt ekstremitelerde akut monoartrit ve kronik mono-oligoartrit olarak görülmektedir. AAA olan hastalarda spondiloartrit nadiren görülmektedir. Burada biz seronegative spondiloartrit ilişkili AAA olan bir vakayı sunuyoruz.

Anahtar Kelimeler

AAA; Bel Ağrısı; Sakroileit; Seronegative Spondiloartrit

Abstract

Familial Mediterranean Fever (FMF) is an autosomal recessive disease characterized by recurrent episodes of febrile serositis. The most frequently seen symptom is abdominal attacks and the second is articular attacks. Joint involvement is usually seen in lower extremities as acute monoarthritis and chronic mono-oligoarthritis. Spondyloarthritis is rarely seen in patients with FMF. Herein we present a case of seronegative spondyloarthritis (SSpA) associated with FMF.

Keywords

FMF; Low Back Pain; Sacroiliitis; Seronegative Spondyloarthritis

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Introduction

Familial Mediterranean Fever (FMF) is an autosomal recessive disease of unknown aetiology which is characterized by recurrent episodes of febrile serositis. Forty-three percent of the patients have familial history indicating a genetic tendency. MEFV gene was found to be related with the development of disease in 77% of the cases [1]. Arthritis and arthralgia in lower extremities are common in FMF patients but spondyloarthritis (SpA) was reported rarely. Despite being a rare condition; sacroiliac joint involvement should be kept in mind when encountered to a FMF patient with low back pain [2]. Herein we present a case of seronegative spondyloarthritis (SSpA) associated with FMF.

Case Report

A 23 years old male patient applied to our clinic with the complaints of inflammatory low back pain for nearly 8 years and intermittent swelling in ankles and knees. He had also attacks of fever, diarrhea and abdominal pain lasting for 1-3 days. These complaints had been proceeding for nearly 8 years. On satisfying criteria for the diagnosis with ongoing complaints, he had been diagnosed with FMF in a rheumatology clinic two years ago and given colchicine 1.5 mg/day. After being initiated colchicine, his attacks decreased in frequency and severity also supporting the diagnosis [3]. His family history was negative with regard to SSpA. The patient did not have a history of uveitis. In physical examination he had prominent growth retardation with 145 cm height and 42 kg weight. He had been investigated in terms of growth retardation in the internal medicine clinic of our hospital 11 months prior to our examination and no explicit reason for growth retardation was found. He was informed that he had genetically short stature. His lumbar and hip movements were in normal range, chest expansion was 4 cm and mennel test was positive bilaterally. In his laboratory analyses; complete blood cell count, urinalysis, liver and kidney function tests were normal. Erythrocyte sedimentation rate (ESR) was 10 mm/h (< 20 mm/h) and C-reactive protein (CRP) was 17.8 mg/dl (0-5 mg/dl). Hepatic markers, HIV tests, serum rheumatoid factor, agglutination test for brucella, urine and throat cultures were negative and purified protein derivative was 1 cm. Abdominal ultrasonography revealed a slight increase in the sizes of liver and spleen. In the light of these findings we focused on gluten-sensitive enteropathy, inflammatory bowel disease and SSpA. In order to clarify diarrhea aetiology, the patient went on colonoscopy which did not reveal any lesions supporting inflammatory bowel disease. Anti-gliadin IgA, anti-endomysial antibody and tissue transglutaminase IgA were negative and his gastrointestinal tract endoscopy was not reveal any lymphoepithelial involvement regarding gluten-sensitive enteropathy. HLA-B27 was negative and he had homozygous M694V mutation in the MEFV gene. Bilateral grade II sacroiliitis was observed on posteroanterior pelvic radiography (Fig. 1). Consequently we diagnosed the patient with FMF-related SpA due to inflammatory back pain ongoing for 8 years, bilateral grade 2 sacroiliitis on posteroanterior pelvic radiography and due to HLA B 27 negativity, absence of significant changes in spinal radiography such as bamboo spine and absence of articular involvement of the anterior thoracic wall and uveitis [4]. Sulfasalazine 2 g/day and diclofenac sodium 100 mg/day in addition to colchicine treatment as well as posture and breathing exercise program was started. He has recovered clinically in his follow up. His CRP values reduced into normal ranges after 7 weeks of treatment.



Figure 1. Postero-anterior pelvic radiography

Discussion

FMF is an inherited genetic disease characterized with recurrent episodes of fever, attacks of peritonitis, pleuritis and synovitis [1]. Joint involvement is the second frequent symptom after the abdominal attacks. Three forms of the arthritis were described in FMF which are acute, chronic and abortive. Acute arthritis occurs in 95% of the reported cases. It usually affects the big joints especially hip, knee and ankle. Chronic arthritis, which may last for more than a month represents in 5% of the cases. The third type is the abortive attack in which arthralgia is the main symptom [2].

Sacroiliac joint involvement is uncommon and HLA-B27 is negative in almost all of these patients [5]. Langevitz et al. reported 11 patients to fulfil the criteria of SSpA in 3000 FMF patients. All of these patients were negative for HLA-B27 [6]. Brodey et al. revealed radiographic sacroiliitis in 6 of 43 FMF patients [7]. In another study, frequency of sacroiliitis among all FMF patients was found to be 7% [1]. The different results of these studies cited above may be due to the distinctness in the patient populations and the radiological methods for detecting sacroiliitis.

In literature; the first case of FMF in association with SSpA was described in 1963 [8]. Thereafter, this association was reported in case reports from either our country or abroad. A 43-years-old male patient with concomitant FMF and psoriasis presenting with bilateral sacroiliitis, chronic hip and knee arthritis in 2008 [9], 3 patients with FMF and SSpA in 2005 [10] and 2 male FMF patients with sacroiliitis in 2009 were also reported [11].

Our patient had inflammatory low back pain, abdominal pain attacks, diarrhea, arthritis and growth retardation in his history. No explanatory reasons for growth retardation was detected. When we screened the literature we could not find any data reporting an increase of SpA frequency among patients with growth retardation. Additionally, our patient's findings satisfied the Assessment in Spondylo Arthritis International Society SSpA criteria [12]. The patient was negative for HLA-B27, did not have uveitis or psoriasis history and his radiographic inspection did not include any signs pointing AS like syndesmoph-

ytes, bamboo spine or squaring of vertebrae. Thereby we diagnosed the patient as FMF-related SSpA. In his follow-up period our patient showed recovery with diclofenac sodium 100 mg/day and sulfasalazine 2 gr/day which were added to his ongoing colchicum 1.5 mg/day treatment. Both his clinical condition and laboratory analysis enhanced with the treatment.

Actually the pathogenetic relationship between FMF and AS remains ambiguous. Further studies to investigate the relationships between these diseases and to determine the appropriate treatments in patients likely to have protracted attacks are warranted. We propose that patients with FMF should be examined with regard to SSpA in case of inflammatory low back pain. On the other side FMF should also be considered in the differential diagnosis of SSpA and rheumatologic diseases with spinal involvement.

Competing interests

The authors declare that they have no competing interests.

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